

·专家论坛·

结缔组织病累及呼吸系统的影像特点分析

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【摘要】结缔组织病累及呼吸系统可表现为间质性肺疾病、弥漫性肺泡损伤、肺泡出血、肺血管病变、胸膜病变及气道病变等。其临床表现缺乏特异性，肺功能检查和胸部 CT 是最常用的检查手段。不同结缔组织病累及呼吸系统的影像特征具有相似性，但又各具特点。本文就结缔组织病呼吸系统受累相关影像表现进行概述，以提升读者的认知并指导临床实践。

【关键词】呼吸系统；结缔组织病；影像学

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Imaging Manifestations of Respiratory Diseases Associated with Connective Tissue Diseases

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【**Abstract**】 Respiratory manifestations of connective tissue diseases include interstitial lung diseases，diffuse alveolar injury，alveolar hemorrhage，pulmonary vascular lesions，pleural lesions and airway disease. Their clinical symptoms were not specific. Pulmonary function test and chest CT come to be the most frequently applied examinations. Different connective tissue diseases involving respiratory system have not only common features but also characteristic imaging patterns. In this paper，imaging manifestations of respiratory diseases associated with connective tissue diseases were reviewed，so as to improve readers' recognition of the diseases and guide clinical practice.

【**Key words**】respiratory system；connective tissue diseases；imaging

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间质性肺疾病（interstitial lung disease，ILD），又称弥漫性实质性肺疾病，指主要累及肺间质、肺泡和（或）细支气管的一组弥漫性肺部疾病，但不包括细支气管以上的各级支气管疾病，可分为已知原因的ILD、肉芽肿性 ILD、罕见但具有临床病理学特征的ILD 及特发性间质性肺炎。

结缔组织病（connective tissue diseases，CTD）易累及呼吸系统的各个器官，包括肺实质、肺血管、气道及胸膜等，表现为 ILD、弥漫性肺泡损伤（diffuse alveolar damage，DAD）、肺泡出血、肺血管病变、胸膜病变及气道病变等，最常见的为 ILD 及肺高血压（pulmonary hypertension，PH）。

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CTD 累及呼吸系统的临床表现缺乏特异性，肺功能检查可发现呼吸动力学改变，胸部增强 CT 和 CT 肺动脉成像（CT pulmonary angiography，CTPA）用于发现肺动脉受累情况，胸部高分辨 CT（high resolution CT，HRCT）可发现肺实质、肺间质及气道的形态结构改变，CT 肺功能检查能够显示肺通气功能变化[1]。文献报道，一氧化碳弥散功能明显下降的 CTD 患者，胸部 HRCT 易出现异常，且 HRCT 显示的病变范围与一氧化碳弥散功能、肺活量、1 秒用力肺活量有一定相关性[2]。因此，肺功能检查与胸部 CT 检查成为评价 CTD 累及呼吸系统最常用的检查手段。本文就 CTD 呼吸系统受累相关影像表现进行概述。

1　结缔组织病相关呼吸系统疾病影像学表现

1.1　肺高血压

PH 定义为安静状态、仰卧位右心导管测量肺动脉平均压≥25 mm Hg（1 mm Hg＝0. 133 kPa），涵盖肺动脉高压及肺纤维化所致 PH[3]。国外报道 CTD 相关 PH 最常见于系统性硬化症（systemic sclerosis，SSc），其次为混合性结缔组织病（mixed connective tissue disease，MCTD）及系统性红斑狼疮（systemic lupus erythematosus，SLE），而中国人多见于 SLE[3-4]。CTD 相关 PH 起病隐匿，患者常死于右心衰竭，肺功能检查示一氧化碳弥散功能下降。胸部增强 CT 或 CTPA 表现为肺动脉及其分支增粗、右心增大、对比剂返流至下腔静脉及肝静脉，严重者出现心包积液。胸部增强 CT 同时可检出 ILD 及肺血栓[5]。郑亚国等[6]报道，以主肺动脉直径 30 mm 为界值，胸部增强 CT 或 CTPA 诊断肺动脉高压的敏感度为 90. 8%，特异度为 66. 7%；以主肺动脉与升主动脉直径比值 1. 0 为界值，诊断肺动脉高压的敏感度为 94. 3%，特异度为 55. 6%，故 CT 能够有效预测肺动脉高压。磁共振成像可作为补充检查手段，评价右心室结构、功能及肺动脉血流[7]。

1.2　间质性肺疾病

胸部 HRCT 可评价 CTD 相关 ILD 的肺内病变是否处于活动期及对治疗的反应[1]。胸部 HRCT 表现为磨玻璃影（ground-glass opacity，GGO）时，多提示 CTD 相关 ILD 的早期肺泡炎，但 Anaya 等[8]认为，GGO 并不一定是急性病变；胸部 HRCT 表现为小叶间隔增厚、粗网状影、蜂窝影，多提示 CTD 相关 ILD 的肺纤维化。非特异性间质性肺炎（nonspecific interstitial pneumonia，NSIP）在 CTD 相关 ILD 中最常见[9-10]，HRCT 表现为双肺下叶胸膜下对称分布的 GGO、细网状影以及支气管扩张，病变程度轻，气腔实变与蜂窝影少见[11]。纤维型 NSIP 还可见肺下叶体积缩小，但细胞型与纤维型 NSIP 从影像上很难鉴别[12]。除 NSIP 外，也可见寻常性间质性肺炎（usual interstitial pneumonia，UIP）、机化性肺炎（organizing pneumonia，OP）、淋巴细胞性间质性肺炎（lymphocytic interstitial pneumonia，LIP）等。虽然 CTD 累及呼吸系统具有一定共性，但每种 CTD 仍各具特点。

2　不同结缔组织病累及呼吸系统的影像特征

2.1　系统性红斑狼疮

胸膜炎是 SLE 最常见的胸部表现，与胸腔积液相关或不相关[1]。SLE 急性肺损伤表现为肺出血、狼疮肺炎，虽临床罕见，但常可致死。慢性肺损伤表现为 ILD[13]。SLE 弥漫性肺泡出血时，患者出现呼吸困难、发热、咳血痰，血色素下降等，HRCT 表现为双肺弥漫或局灶实变影、GGO 及小叶间隔增厚，此时应行支气管镜检查以确诊[14]。急性狼疮肺炎的诊断难以界定，表现为不同程度肺功能受损，HRCT 示弥漫或散在分布的气腔实变与 GGO，下肺相对较重，半数患者合并胸腔积液[15]。但目前认为，既往诊断为急性狼疮肺炎的病变很可能为急性间质性肺炎，伴或不伴肺出血[1]。仅 3% 的 SLE 患者中出现 ILD，以 NSIP 或 UIP 最多见，SLE 相关 ILD 的病变常局限、程度较轻，弥漫性病变、蜂窝影很少出现，多表现为小叶间隔增厚或肺实质带[16]。活动期的 SLE 患者可出现深静脉血栓，若患者出现胸痛、呼吸困难，需行胸部增强 CT 或 CTPA 以明确是否存在肺血栓、肺梗死[17]。SLE 患者胸部 HRCT 表现为 GGO 或肺实变，可能为 ILD、肺出血、狼疮肺炎等，若同时出现小叶间隔增厚、不规则线影、肺结构变形，意味着肺纤维化，多见于 ILD 等[18-20]。SLE 易合并感染，多数为社区获得性肺炎，诊断 ILD、急性狼疮肺炎、肺泡出血等需先除外感染[21]。另外，SLE 患者还可出现横膈功能障碍、肺动脉高压等。

2.2　类风湿关节炎

类风湿关节炎（rheumatoid arthritis，RA）累及胸部常出现胸膜病变、气道疾病（支气管扩张、滤泡性支气管炎、闭塞性支气管炎等）、ILD、RA 结节等[22-23]。RA 胸膜病变 HRCT 表现为轻度胸膜增厚，少量、单侧的胸腔积液，胸腔积液多为自限性，不一定合并肺内病变[24]。气道病变通常为 RA 肺内最早出

现的征象，约30%～58%的RA患者HRCT可见支气管扩张[25]。闭塞性细支气管炎HRCT表现为马赛克征、小气道闭塞、支气管扩张，深呼气相HRCT可见空气潴留；滤泡性细支气管炎HRCT多表现为磨玻璃样密度的小叶中心结节[26-27]。ILD仅见于5%的RA患者[25]，类风湿因子阳性、关节损害重者易出现ILD[28]。RA相关ILD的HRCT多表现为网状影（小叶间隔增厚、小叶内间质增厚等）、蜂窝影伴支气管扩张，GGO范围小且多分布于肺底部，进展期可见肺体积缩小及肺结构变形[23,29]（图1）。HRCT随诊观察发现肺小叶中心线状影易进展为支气管扩张，气腔实变易进展为蜂窝[28]。单纯RA相关ILD以UIP多见，合并其他CTD则以NSIP最多见[30]。RA肺结节常见于吸烟的男性RA患者，患者常同时伴有皮下结节，HRCT表现为肺结节多位于中上肺周边，大小不等，类圆形，边界清，可伴空洞，结节新发与自限并存，钙化少见，仅见于煤工尘肺的RA患者[31-32]。

2.3 系统性硬化症

SSc较其他CTD更易累及肺，80%患者可出现ILD，以NSIP最多见，UIP也可见[33-35]。SSc相关ILD的HRCT表现为位于肺下叶、周边的网状影、蜂窝影及GGO，背侧最重（图2）；可伴有支气管扩张、黏液栓、小叶中心结节及小斑片实变，胸腔积液相对少见[36]。HRCT随诊中，蜂窝影与GGO多逐渐加重，GGO进展程度与一氧化碳弥散功能呈显著负相关；若治疗后GGO不可逆，则GGO病理上并非炎症，而是纤维化[37-39]。SSc累及食道易出现吸入性肺炎或支气管炎，累及肺血管可出现持续肺动脉高压[34-35,40]。进行性系统性硬化症易合并肿瘤，以肺癌、乳腺癌最多见[41]，需注意随诊筛查。

2.4 多肌炎/皮肌炎

多肌炎/皮肌炎（polymyositis/dermatomyositis，PM/DM）累及呼吸肌可致肺通气功能下降，亦可出现吸入性肺炎[42]。患者抗J0-1抗体阳性与ILD明显相关，约50%～70%的抗J0-1抗体阳性PM/DM患者出现ILD，而阴性者伴ILD仅占10%[1]。ILD病理上以OP和NSIP最多见，二者可同时存在[43-45]（图3）。PM/DM相关ILD的胸部HRCT多表现为GGO、斑片状气腔实变、肺实质带、小叶间隔增厚、胸膜下线、支气管血管束增粗、支气管扩张等。斑片状气腔实变伴或不伴GGO，多为OP或机化的DAD；治疗后气腔实变、实质带、不规则支气管血管束增粗多可逆，局灶GGO伴实质带或胸膜下线则可进展为蜂窝影[46-48]。PM/DM患者易出现肿瘤，特别是肺癌，胸部CT随诊过程中新出现肺结节，需注意鉴别。



图1 男性，66岁，类风湿关节炎，高分辨CT示双肺多发磨玻璃密度影、网状影、蜂窝影及支气管扩张，诊断为寻常性间质性肺炎

2.5 干燥综合征

干燥综合征（Sjögren’s syndrome，SS）相关ILD以LIP、NSIP多见，累及气道可出现支气管扩张、滤泡性细支气管炎、细支气管炎等，另外可见肺动脉高压、淋巴瘤、胸膜病变等，且均多见于下肺[49-52]。50%的LIP患者HRCT表现为GGO伴散在的囊状气腔（图4），可伴有小叶中心、支气管血管束周围、胸膜下结节及支气管血管束增粗，蜂窝与实变少见[29,47,53-54]。

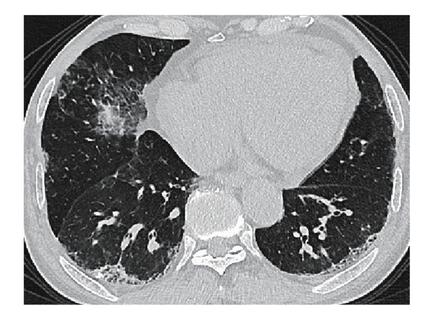


图2 男性，66岁，系统性硬化症，高分辨CT示双肺胸膜下多发磨玻璃密度影及网状影，右肺中叶多发支气管扩张，诊断为非特异性间质性肺炎



图3 女性，43岁，皮肌炎，高分辨CT示双肺近胸膜下多发实变影、磨玻璃密度影、小叶间隔增厚及支气管扩张，诊断为机化性肺炎和非特异性间质性肺炎

囊腔影有助于鉴别 LIP 与淋巴瘤。SS 易合并淋巴瘤，肺部以黏膜相关淋巴组织淋巴瘤为多，当肺内出现实变、结节大于 1cm 或胸腔积液时，需考虑到淋巴瘤的可能[55-56]。

2.6 混合性结缔组织病

MCTD 是一种以 SLE、SSc、PM/DM 及 RA 样症状重叠为特征的特殊病变类型，有极高滴度的循环抗核抗体。MCTD 累及胸部以肺动脉高压、ILD、胸膜增厚、胸腔积液、心包积液最多见[57]。MCTD 相关 ILD 主要表现为 NSIP，HRCT 多表现为双下肺胸膜下 GGO（图 5），蜂窝、实变、小叶中心结节等较少见[9,24,58]。

2.7 强直性脊柱炎

强直性脊柱炎肺部受累多见于 AS 病程较长的患者，HRCT 示病变主要位于双上肺，表现为肺纤维化、囊状气腔、空洞、支气管扩张、间隔旁型肺气肿及肺实质带等，有时与肺结核鉴别困难[36,59-60]。



图 4 女性，47 岁，干燥综合征，高分辨 CT 示双肺多发薄壁囊腔影及小叶中心结节，诊断为淋巴细胞性间质性肺炎

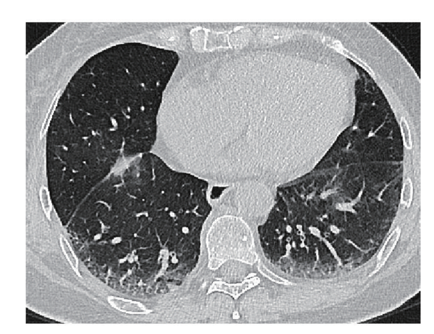


图 5 女性，53 岁，混合性结缔组织病，高分辨 CT 示双肺下叶胸膜下多发磨玻璃密度影、小叶间隔增厚，诊断为非特异性间质性肺炎

3 小结

胸部 HRCT 为评估 CTD 病变的首选检查，其能够及时、准确发现 CTD 呼吸系统受累的异常征象，有助于评估病变累及范围、程度、治疗效果及预后。临床上，CTD 系统性症状不明显时，仅凭 HRCT 很难鉴别 CTD 相关 ILD 与特发性 ILD，需结合自身抗体等实验室检查综合判断。另外，在诊断及评价 CTD 肺部受累时，需结合临床表现、治疗情况及实验室检查等综合考虑，首先排除药物治疗相关的并发症及机遇性肺炎所致病变，再行评估[61]。CTD 病变影像表现多样，且不同 CTD 形态学征象存在重叠，在评价可疑 CTD 患者时，抓住 CTD 病变的特殊胸部影像学特点，有助于理清思路，最大程度获取诊断信息。此外，系统、全面地评价患者胸部病变（肺实质、气道、胸膜、肺血管等），有利于临床鉴别诊断、获得准确的评估。

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